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# Relationship between initial electroencephalographic characteristics and seizure outcomes in children with non-lesional West syndrome

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**Summary**
*Background:* To characterize the initial interictal electroencephalography (EEG) activities associated with seizure outcomes in children with non-lesional West syndrome (WS), and their worth in the prediction of seizure-free (SF) vs no seizure-free (No-SF) outcomes.
*Methods:* We retrospectively reviewed the initial scalp EEGs for at least a 40-min duration, and the medical records of 66 children who were diagnosed as WS with normal MRI, and who were followed-up with for 4.5 ± 2.1 years. We assessed the following clinical and EEG findings: onset of seizures, development, underlying etiologies, initial interictal EEGs, and seizure evolution. These variables were compared between two groups: SF vs No-SF groups.
*Results:* In total, 36 (54.5%) children had SF outcomes and 30 (45.5%) had No-SF outcomes during long-term follow-up (4.4 ± 2.3 vs 4.6 ± 2.0 years, $p = 0.7644$). The mean age at seizure onset

*Abbreviations:* WS, West syndrome; PET, positron emission tomography; EEG, electroencephalogram; ED, epileptiform discharges; AEDs, antiepileptic drugs; MISF, multifocal independent spike foci; SF, seizure-free; No-SF, no seizure-free.

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was similar in the SF and No-SF groups (6.0 ± 3.0 vs 6.9 ± 3.2 months, $p = 0.2443$). Delayed development before the onset of spasms was similarly observed in both groups (13.9% vs 13.3%). Initial EEG findings significantly differed with typical hypsarrhythmia (41.7% vs 73.3%, $p = 0.0098$), multiple independent spike foci (MISF) (55.6% vs 83.3%, $p = 0.0158$), frontal-dominant MISF (0.0% vs 40.0%, $p < 0.0001$), and frontal-dominant generalized epileptiform discharges (EDs) (0.0% vs 16.7%, $p = 0.0108$) being involved more infrequently in the SF group than in the No-SF group, respectively. Patients in the SF group showed no frontal-dominant MISF or frontal-dominant generalized EDs, and a more often normal to borderline sleep-spindle (83.3% vs 40.0%, $p = 0.0002$) than the No-SF group.

*Conclusion:* Patients with SF outcomes more frequently showed the posterior-dominant generalized EDs and normal to borderline sleep-spindle, and the No-SF group more often had typical hypsarrhythmia, frontal-dominant MISF, frontal-dominant generalized EDs, and no normal sleep-spindle. Initial interictal EEG findings may predict seizure outcomes in patients with non-lesional WS.



## Introduction

Despite heterogeneous causes, West syndrome (WS) is an age-dependent disorder, often occurring at 3—8 months of age (Yamatogi and Ohtahara, 1981; Ohtahara, 1984), and it is characterized by epileptic spasms and hypsarrhythmia. It is typically associated with developmental arrest or regression, as well as poor seizure outcomes (Watanabe, 1998; Hamano et al., 2003; Riikonen, 2004). WS is usually classified into cryptogenic or symptomatic origins. Outcomes associated with cryptogenic WS are usually more favorable than those of the symptomatic group (Koo et al., 1993; Nabbout, 2001; Ito et al., 2002; Hamano et al., 2003, 2006). However, almost half of those cases have moderate to severe mental retardation (Koo et al., 1993; Ito et al., 2002; Hamano et al., 2003). Cryptogenic WS is considered to be 'probably symptomatic', or more precisely, to have unidentified underlying biochemical or structural causes. Nevertheless, one researcher has recognized the continuation of idiopathic cases (Lux and Osborne, 2004). The diversities of underlying causes are probably associated with multifarious outcomes of cryptogenic WS.

In a study of the variables affecting the developmental outcome of cryptogenic WS (Hamano et al., 2007), it was suggested that a shorter treatment lag may be associated with a favorable outcome, and that paroxysmal discharges in the frontal area and symptom evolution to other seizure types may be related to unidentified lesions there. Another study revealed that the hypometabolism of positron emission tomography (PET) after initial pharmacologic treatment was correlated with the poor seizures and developmental outcomes (Natsume et al., 2014). PET could detect subtle cortical abnormalities in the patients considered to be cryptogenic because of normal brain images, and a pathological examination showed that resected tissues corresponding to the hypometabolic area were malformative and dysplastic cortical lesions (Chugani et al., 1993). However, the number of institutions equipped with PET facilities is limited; the majority of the institutions have the facilities for electroencephalographic (EEG) recording, which is much more common and convenient.

The relationship between the distinctive features of interictal epileptiform discharges (EDs) on initial scalp EEGs and seizure outcomes in patients with WS with normal MRI has remained uncertain. The aim of this study is to evaluate and analyze the characteristics of initial interictal EEG activities in children with non-lesional WS, and to assess its value in predicting favorable or poor seizure outcomes.

## Patients and methods

### Patients

We retrospectively investigated those children with non-lesional WS at the Department of Pediatric Neurology, Pusan National University Children's Hospital and Pusan National University Hospital from 2002 to 2010, who were examined regularly for more than 1 year by a pediatric neurologist. Non-lesional WS was defined as follows: (a) clusters of epileptic spasms with onset <1 year after birth, (b) typical/classic or modified hypsarrhythmia (refer to Section ''Methods'') including generalized or multifocal EDs on interictal EEG before treatment with antiepileptic drugs (AEDs), (c) normal birth and absence of any etiologic factors related to WS, (d) no eventful past history including no other types of seizures before onset of spasms, (e) no focal neurologic abnormalities at the onset, and (f) normal brain images on MRI. We excluded patients with (a) abnormal fetal life or remarkable perinatal problems, (b) known chromosomal anomalies or inborn errors of metabolism, (c) a dysmorphic face or congenital anomaly of other organ, (d) an absence of initial EEG recordings prior to the start of AEDs, and (e) the use of a sedative drug for EEG recording. In total, 66 patients (39 boys and 27 girls) with non-lesional WS were selected to participate in this study.

### Methods

EEGs were recorded using a Grass-Telefactor system (Telefactor Cop, Conshohoken, PA) or a Stellate Harmonie EEG (Stellate System Inc., Canada) with a high-cut filter at 70 Hz for more than 40 min during sleep, but with no fully alert period. All patients slept spontaneous for the